

Eun Young Kim and Yeon Hyeon Choe

Contents

13.1	Overview	175
13.1.1	Definition	175
13.1.2	Prevalence	175
13.1.3	Clinical Features	176
13.1.4	Cause	176
13.2	Imaging Modalities and Findings	176
13.2.1	Computed Tomography	176
13.2.2	Magnetic Resonance Imaging	176
13.3	Summary	180
	References	180

Abstract

Dilated cardiomyopathy (DCM) is a progressive disease of heart muscle that is characterized by ventricular chamber enlargement and contractile dysfunction, and DCM is the third most common cause of heart failure and the most frequent reason for heart transplantation. Cardiac MR is useful modality for the diagnosis, and to assess the degree of cardiac dysfunction, to identify the cause, and to guide treatment.

13.1 Overview**13.1.1 Definition**

- Ventricular chamber enlargement and systolic dysfunction (left ventricular ejection fraction <30–40 % or fractional shortening less than 25 %) [1, 2].

13.1.2 Prevalence

- Five to eight cases per 100,000 populations, with an estimated prevalence of 1:2,500 [3].
- The third most common cause of heart failure after ischemia and valvular disease.
- Approximately 90 % of all cardiomyopathies; approximately 50 % of all cases of dilated cardiomyopathy (DCM) are idiopathic [4].
- Idiopathic DCM is the most common cause of heart failure in the young, with an estimated prevalence of at least 36.5 per 100,000 persons in the United States.
- Due to mild clinical symptoms in the early phase of the disease, the true prevalence is probably even much higher. It has been suggested that up to 14 % of the middle-aged and elderly population have asymptomatic left ventricular systolic dysfunction [5].

Electronic supplementary material Supplementary material is available in the online version of this chapter at [10.1007/978-3-642-36397-9_13](https://doi.org/10.1007/978-3-642-36397-9_13).

E.Y. Kim

Department of Radiology, Gachon University
Gil Hospital, Incheon, Republic of Korea
e-mail: oneshot0229@gmail.com

Y.H. Choe (✉)

Department of Radiology, Samsung Medical Center,
Sungkyunkwan University School of Medicine,
Seoul, Republic of Korea
e-mail: yhchoe@skku.edu

Table 13.1 Causes of dilated cardiomyopathy

Ischemia	Medications	Rheumatologic disease
Infection	Chemotherapeutic agents	Systemic lupus, scleroderma
Virus	Antiretroviral drugs	Endocrinologic disorders
Bacteria	Phenothiazines, chloroquine	Pheochromocytoma, diabetes mellitus
Fungus	Electrolyte abnormalities	Miscellaneous
Parasite	Hypocalcemia, uremia	Radiation
Rickettsia	Hypophosphatemia	Sarcoidosis
Deposition disease	Genetic ± neuromuscular disease	Tachycardia
Hemochromatosis	Duchenne's muscular dystrophy	Sleep apnea
Amyloidosis	Myotonic dystrophy	Oxygen free radical
Toxins	Friedreich's ataxia	Autoimmune myocarditis
Ethanol, cocaine	Nutritional deficiencies	Familial cardiomyopathies
Lead, mercury	Thiamine, selenium, carnitine	Peripartum cardiomyopathy

13.1.3 Clinical Features

- Most commonly diagnosed in the third or fourth decade, but also in young children [3].
- Progressive heart failure and a decline in left ventricular systolic function, arrhythmias, thromboembolism, and sudden death at any stage of the disease.
- High mortality rate (median period of survival of 1.7 years for men and 3.2 years for women) [3].
- The natural history of the condition is progressive, and its cost, disability, and morbidity are among the highest of any disease.
- Histopathologic features – generally microscopic interstitial fibrosis, but some patients have grossly visible nontransmural or, rarely, transmural fibrosis [6].
- Systolic dysfunction is the most important independent predictor of outcome, and evaluation of diastolic filling allows further identification of subgroups with divergent long-term prognosis.

13.1.4 Cause (Table 13.1)

- In the World Health Organization classification, DCM is classified as its primary (e.g., idiopathic or familial) and secondary forms.
- Up to 50 % of patients diagnosed with idiopathic cardiomyopathy have a familial DCM.
- Although genetically heterogeneous, the predominant mode of inheritance for DCM is autosomal dominant, with X-linked autosomal recessive and mitochondrial inheritance less frequently.

13.2 Imaging Modalities and Findings

13.2.1 Computed Tomography

- With ECG-gated cardiac CT, coronary artery disease can be excluded because of high specificity and negative predictive value.

- Although ionizing radiation and injection of relatively large amounts of iodinated contrast agents are required, ECG-gated CT scanning enables morphological analysis of the ventricles and is an accurate means of evaluating ventricular function (Fig. 13.1).

13.2.2 Magnetic Resonance Imaging

- Detailed morphologic evaluation of ventricles.
 - In black blood images, enlarged cardiac chambers and thin myocardial walls are evident.
 - Mural thrombi can also be identified.
- Functional evaluation of ventricles.
 - Cine images usually show ventricular hypokinesia and increased volumes. Using steady-state free precession (SSFP) images, the diagnosis of left ventricle (LV) dilation is simply made when short-axis internal LV chamber diameter is larger than 5.0 cm or when the LV end diastolic volume exceeds 235 mL or 112 mL/m² in males and 174 or 99 mL/m² in females.
 - The superior quality of images obtained by SSFP technique facilitates the detection of regional wall motion abnormalities allowing an easier differentiation between ischemic and non-ischemic LV impairment [7].
 - CMR is able to overcome many of the limitations of echocardiographic assessment of ventricular function and volumes. The significantly lower inter- and intraobserver variability in CMR measurements allows better monitoring of response to medical intervention or disease progression.
- Characterization of myocardial tissue using late gadolinium enhancement (LGE) images.
 - To differentiate between DCM secondary to coronary artery disease and other causes of DCM. The differentiation between these subgroups may be fundamental in the therapeutic and prognostic approach to the patients [8].
 - In non-ischemic DCM, hyperenhancement was either absent (59–88 % of cases) or appeared as



Fig. 13.1 CT of a patient with idiopathic dilated cardiomyopathy. ECG-gated cardiac CT shows a dilated left ventricle (7 cm in the internal diameter)

stripes of hyperenhancement in the mid-wall of the myocardium not related to specific coronary artery perfusion territories (9–35 % of the cases).

- A subgroup of patients with DCM has fibrosis in a predominantly subendocardial distribution, characteristic of infarction (it has been suggested that these may represent coronary emboli-induced ischemic cardiomyopathy cases or ruptured coronary plaques that have subsequently recanalized).
- Degree of fibrosis is an important prognostic predictor.
 - In a group of patients with DCM, 35 % of these patients had mid-wall myocardial fibrosis, which is a predictor of the combined end point of all-cause mortality and cardiovascular hospitalization and also of sudden cardiac death and ventricular tachycardia [9].
 - The predictive value of mid-wall fibrosis remained significant after correction for LV volumes and ejection fraction (Figs. 13.2, 13.3, 13.4, 13.5, and 13.6).

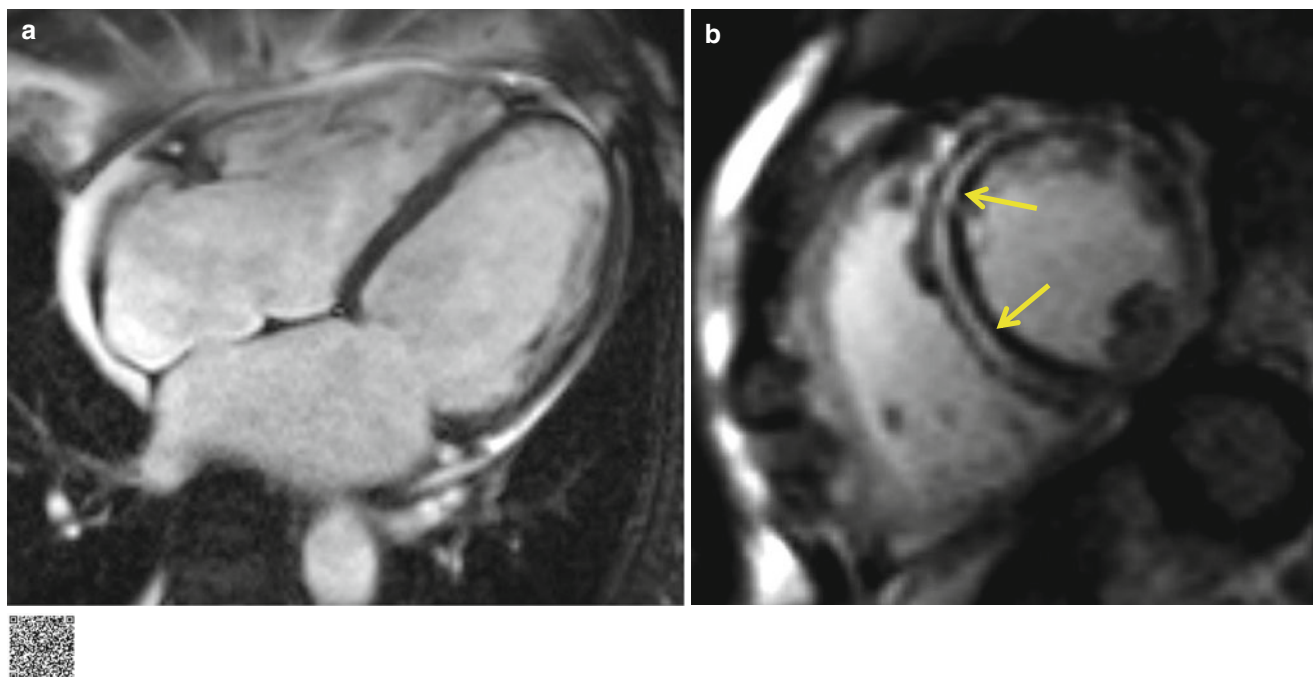


Fig. 13.2 MRI of a patient with idiopathic dilated cardiomyopathy (DCM) (<http://extras.springer.com/2015/978-3-642-36396-2>). (a) Four-chamber cine MRI shows dilated ventricles. Calculated left ventricular ejection fraction using cine MRI was 39 %. (b) Delayed

enhancement MRI demonstrates typical non-ischemic DCM of delayed enhancement (*arrows*) in the LV, i.e., stripes of hyperenhancement in the mid-wall of the myocardium

Learning Points of DCM

Stripes of hyperenhancement in the mid-wall of the myocardium are a typical enhancement pattern in patients with non-ischemic DCM.

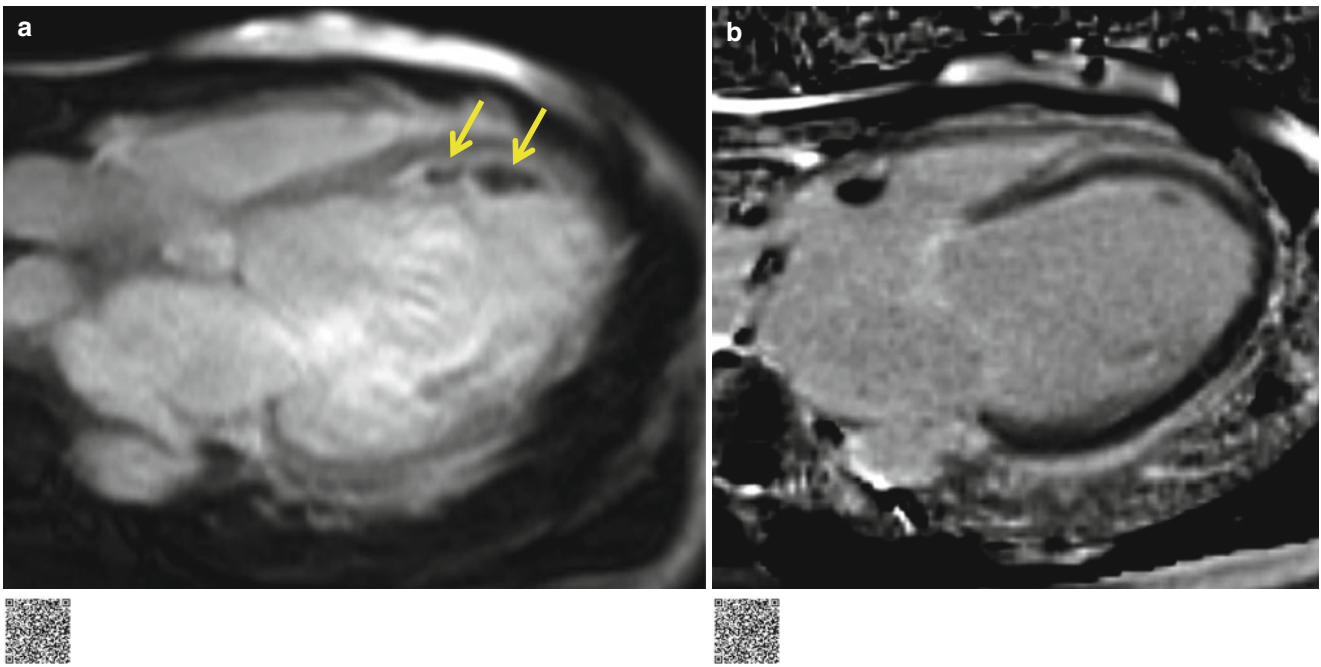
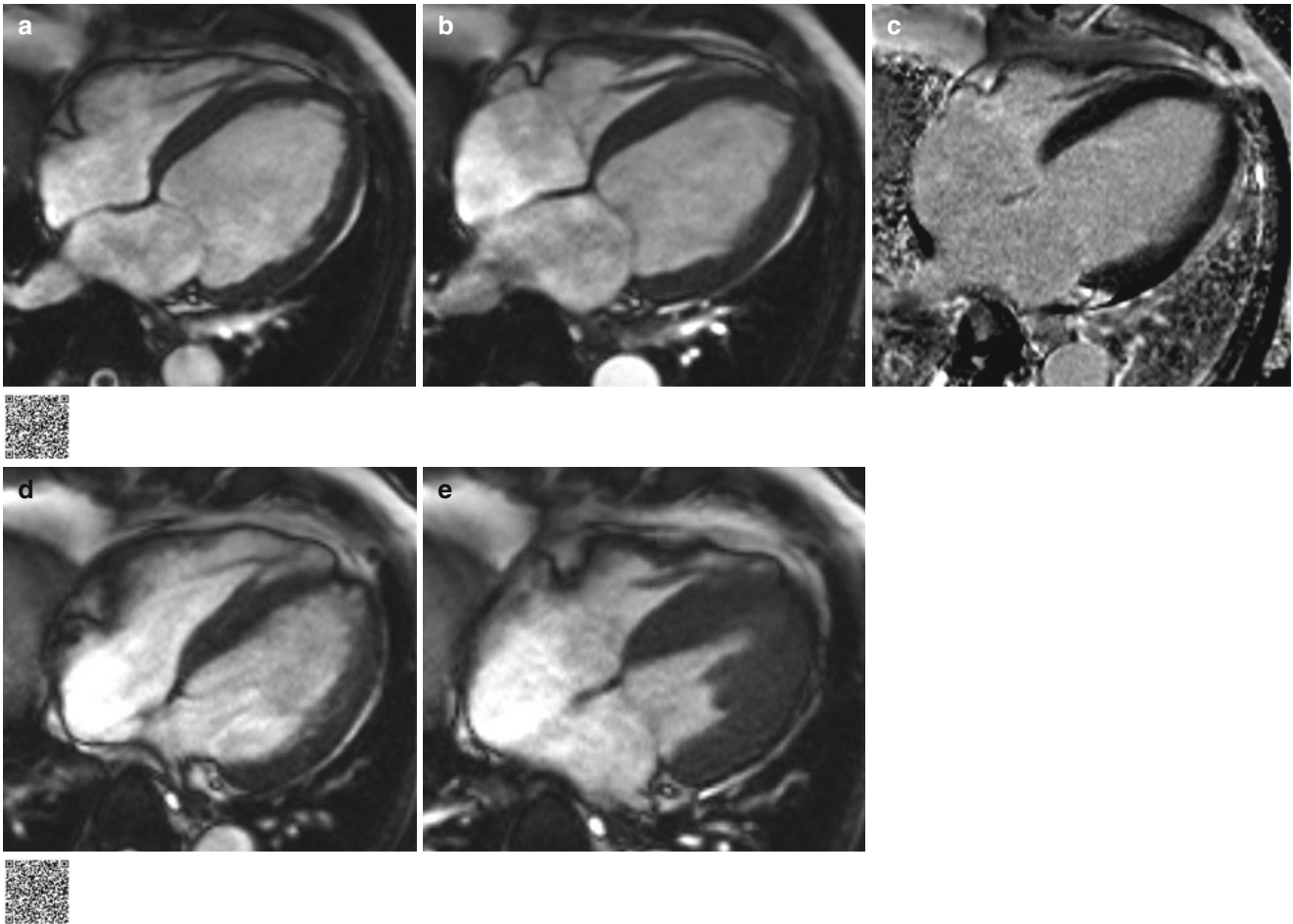


Fig. 13.3 MRI of a patient with idiopathic dilated cardiomyopathy and thrombus in the left ventricle (<http://extras.springer.com/2015/978-3-642-36396-2>). (a) Delayed enhancement MRI with long inversion time (600 ms) demonstrates non-enhancing low signal intensity area (*arrows*),

indicating thrombus in the left ventricle. (b) Delayed enhancement MRI (phase-sensitive inversion recovery) shows no abnormal delayed myocardial enhancement



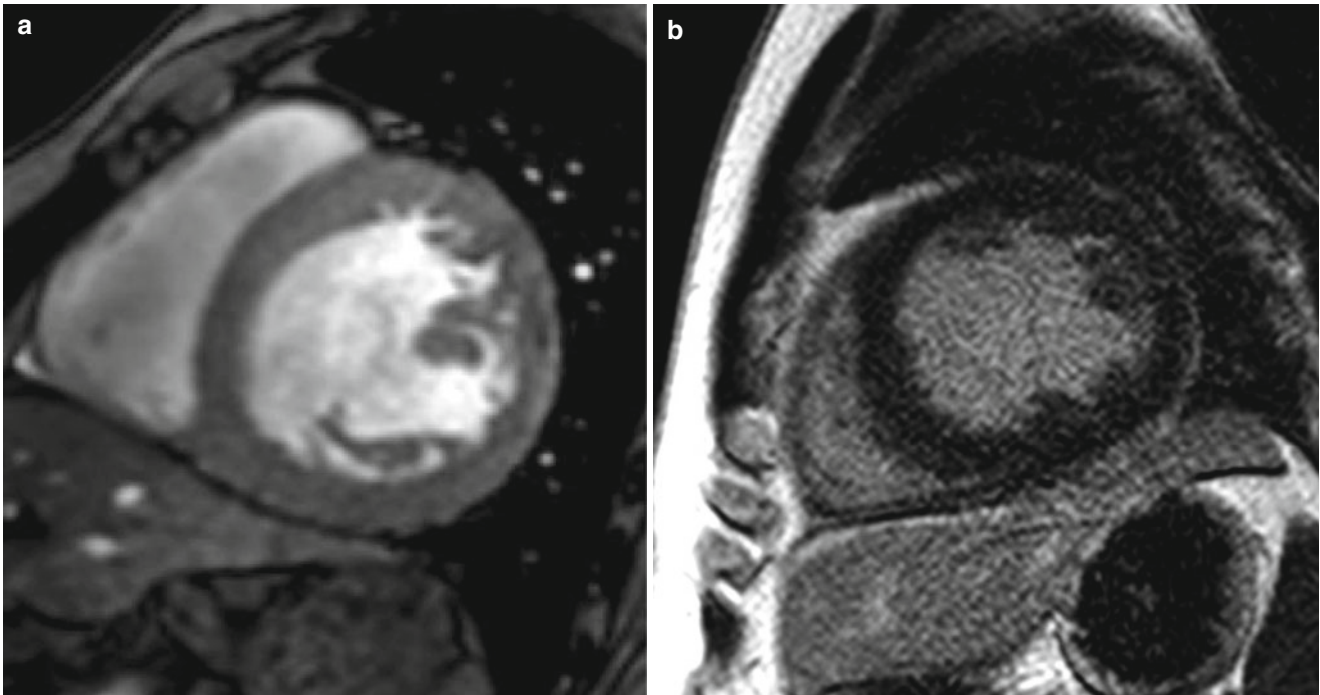


Fig. 13.5 MRI of a patient with a history of excessive alcohol consumption. Invasive coronary angiographic findings were normal (not shown here). (a) Short-axis cine MRI shows a dilated left ventricle.

(b) Delayed enhancement MRI demonstrates no abnormal delayed myocardial enhancement

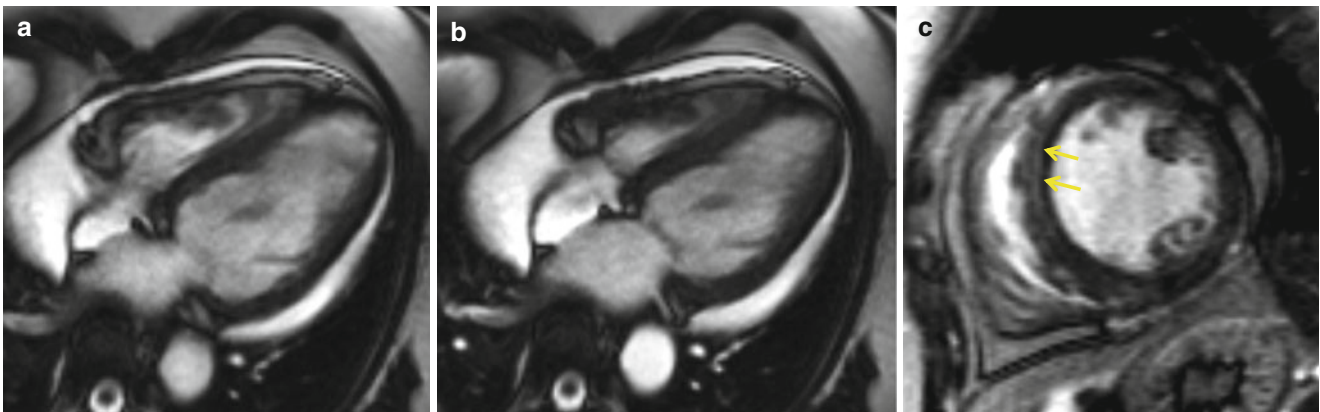


Fig. 13.6 MRI of a female patient with long-term treatment of doxorubicin for malignancy (<http://extras.springer.com/2015/978-3-642-36396-2>). (a) Four-chamber cine MRI shows a dilated left ventricle and impaired systolic contraction of the left ventricle (b systolic phase). Calculated left

ventricular ejection fraction using cine MRI was 23 %, and the left ventricular end diastolic volume was 120 mL/m². (b) Delayed enhancement MRI demonstrates mild mid-wall enhancement in the mid-ventricular septum (arrows)

Fig. 13.4 MRI of a patient with idiopathic dilated cardiomyopathy. (a, b) Initial four-chamber cine MRI shows dilated ventricles and impaired systolic contraction of the left ventricle (b systolic phase) (<http://extras.springer.com/2015/978-3-642-36396-2>). (c) Delayed

enhancement MRI demonstrates no abnormal delayed myocardial enhancement. (d, e). One-year follow-up four-chamber cine MRI reveals normal left ventricular internal dimension and improved systolic contraction (e) (<http://extras.springer.com/2015/978-3-642-36396-2>)

Learning Points of DCM

In non-ischemic DCM, hyperenhancement was either absent (59–88 % of cases) or appeared as stripes of hyperenhancement in the mid-wall of the myocardium (9–35 % of the cases). The myocardial fibrosis (enhancement area) appears to be irreversible and is regarded as a predictor of adverse outcome.

- T1 mapping
 - Postcontrast myocardial T1 time is inversely correlated with the presence of diffuse fibrosis at endomyocardial biopsy in a population with a broad spectrum of cardiomyopathies.
 - Increased gadolinium concentration in the expanded extracellular space associated with scar tissue causes T1 shortening and high signal intensity on T1-weighted images relative to areas of normal myocardium.
 - Significant myocardial fibrosis can be present at endomyocardial biopsy even when cardiac MR images do not show focal LGE. Relatively dense myocardial scar is thought to be necessary for visual identification of myocardial scar with gadolinium-enhanced cardiac MR because of the relatively low resolution of MR imaging [10].
 - In the setting of less severe or more diffuse fibrosis, the inversion-recovery cardiac MR technique is unlikely to reveal the presence of diffusely abnormal tissue given the lack of normal myocardium as a reference.
 - Direct measurement of myocardial T1 time (“T1 mapping”) may improve on these problems of LGE cardiac MR in the setting of more subtle degree of diffuse fibrosis (i.e., DCM, hypertrophic cardiomyopathy, aortic valve disease, postoperative cardiac transplantation, myocarditis, restrictive cardiomyopathy, suspected arrhythmogenic right ventricle dysplasia) [10].

13.3 Summary

- DCM is associated with dilatation and dysfunction of the LV or of both ventricles.
- DCM is caused by a variety of disorders (ischemia, infections, drugs, deposition disease, toxins, electrolyte abnormalities, nutritional deficiencies, endocrine dysfunction, and genetic), although frequently no etiology can be found and the cardiomyopathy is deemed idiopathic.

- CT and MR are used to help make a diagnosis, to assess the degree of cardiac dysfunction, to identify a cause, and to guide therapy.
- Stripes of hyperenhancement in the mid-wall of the myocardium are a typical enhancement pattern, which was identified in a 9–35 % of the patients with non-ischemic DCM, which is a predictor of poor prognosis.

References

1. Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation*. 2006;113:1807–16.
2. Richardson P, McKenna W, Bristow M, et al. Report of the 1995 World Health Organization/International Society and Federation of Cardiology Task Force on the Definition and Classification of cardiomyopathies. *Circulation*. 1996;93:841–2.
3. Dec GW, Fuster V. Idiopathic dilated cardiomyopathy. *N Engl J Med*. 1994;331:1564–75.
4. McDonagh TA, Morrison CE, Lawrence A, et al. Symptomatic and asymptomatic left-ventricular systolic dysfunction in an urban population. *Lancet*. 1997;350:829–33.
5. Devereux RB, Roman MJ, Paranicas M, et al. A population-based assessment of left ventricular systolic dysfunction in middle-aged and older adults: the Strong Heart Study. *Am Heart J*. 2001;141:439–46.
6. Giesbrandt KJ, Bolan CW, Shapiro BP, Edwards WD, Mergo PJ. Diffuse diseases of the myocardium: MRI-pathologic review of cardiomyopathies with dilatation. *AJR Am J Roentgenol*. 2013;200:W274–82.
7. O'Donnell DH, Abbara S, Chaithiraphan V, et al. Cardiac MR imaging of nonischemic cardiomyopathies: imaging protocols and spectra of appearances. *Radiology*. 2012;262:403–22.
8. Belloni E, De Cobelli F, Esposito A, et al. MRI of cardiomyopathy. *AJR Am J Roentgenol*. 2008;191:1702–10.
9. Assomull RG, Prasad SK, Lyne J, et al. Cardiovascular magnetic resonance, fibrosis, and prognosis in dilated cardiomyopathy. *J Am Coll Cardiol*. 2006;48:1977–85.
10. Sibley CT, Noureldin RA, Gai N, et al. T1 Mapping in cardiomyopathy at cardiac MR: comparison with endomyocardial biopsy. *Radiology*. 2012;265:724–32.